

# IgG4-RD Responder Index Validation Study

## Scoring Rules

Scoring refers to manifestations of disease activity present in the last 28 days

- Scoring: 0 Normal or resolved  
 1 Improved but still present  
 2 Persistent (still active; unchanged from previous visit)  
 3 New / Recurrence while patient is off treatment  
 4 Worsened or new disease manifestation despite treatment

## Definitions

**Organ/Site score:** The overall level of IgG4-RD activity within a specific organ system

**Symptomatic:** Is the disease manifestation in a particular organ system symptomatic? (Y = yes; N = no)

**Urgent disease:** Disease that requires treatment immediately to prevent serious organ dysfunction (Y = yes; N = no)  
 (Presence of **urgent disease** within an organ leads to **DOUBLING** of that organ system score)

**Damage:** Organ dysfunction that has occurred as a result of IgG4-RD and is considered permanent (Y = yes; N = no)

| Organ/Site  | Activity               |                      |                 | Damage |                      |
|---|------------------------|----------------------|-----------------|--------|----------------------|
|   | Organ/Site Score (0-4) | Symptomatic (Yes/No) | Urgent (Yes/No) | Yes/No | Symptomatic (Yes/No) |
| Meninges  |                        |                      |                 |        |                      |
| Pituitary Gland   |                        |                      |                 |        |                      |
| Orbital lesion (specify location):<br>_____   |                        |                      |                 |        |                      |
| Lacrimal Glands   |                        |                      |                 |        |                      |
| Parotid Glands  |                        |                      |                 |        |                      |
| Submandibular Glands  |                        |                      |                 |        |                      |
| Other Salivary Glands (specify):<br>_____   |                        |                      |                 |        |                      |
| Mastoiditis / Middle ear disease  |                        |                      |                 |        |                      |
| Nasal Cavity Lesions  |                        |                      |                 |        |                      |
| Sinusitis   |                        |                      |                 |        |                      |
| Other ENT Lesions, e.g., tonsillitis, pharyngitis (specify):<br>_____   |                        |                      |                 |        |                      |
| Thyroid   |                        |                      |                 |        |                      |
| Lungs   |                        |                      |                 |        |                      |
| Lymph Nodes (please circle site of involvement, below):   |                        |                      |                 |        |                      |
| Submental   Submandibular   Cervical   Axillary   Mediastinal   Hilar<br>Abdominal/Pelvic   Inguinal   Other lymph node chains: |                        |                      |                 |        |                      |

| Organ/Site  | Activity               |                      |                    | Damage             |                      |
|---|------------------------|----------------------|--------------------|--------------------|----------------------|
|   | Organ/Site Score (0-4) | Symptomatic (Yes/No) | Urgent (Yes/No)    | Yes/No             | Symptomatic (Yes/No) |
| Aorta / Large Blood Vessels   |                        |                      |                    |                    |                      |
| Heart/Pericardium   |                        |                      |                    |                    |                      |
| Retroperitoneal Fibrosis  |                        |                      |                    |                    |                      |
| Sclerosing Mediastinitis  |                        |                      |                    |                    |                      |
| Sclerosing Mesenteritis   |                        |                      |                    |                    |                      |
| Pancreas  |                        |                      |                    |                    |                      |
| Liver   |                        |                      |                    |                    |                      |
| Bile ducts  |                        |                      |                    |                    |                      |
| Kidney  |                        |                      |                    |                    |                      |
| Skin  |                        |                      |                    |                    |                      |
| Constitutional symptoms not attributable to involvement of a particular organ (weight loss, fever, fatigue caused by active IgG4-RD)                                    |                        |                      |                    |                    |                      |
| Other involvement - specify:<br><br>(Consider prostate, breast, gallbladder involvement; and other. Each "Other" item is counted separately.)<br><br>_____<br><br>_____ | _____<br><br>_____     | _____<br><br>_____   | _____<br><br>_____ | _____<br><br>_____ | _____<br><br>_____   |

### Total Activity Score

Organ/sites (x 2 if urgent): \_\_\_\_\_

Total **urgent** organs: \_\_\_\_\_

Total **symptomatic (active)** organs: \_\_\_\_\_

Total **damaged** organs: \_\_\_\_\_

Total **symptomatic (damage)** organs: \_\_\_\_\_

### Physician Global Assessment (PGA) of disease activity

0

100 mm

PGA = \_\_\_\_\_ mm